Rupture of the abdominal aorta in a 13-year-old girl secondary to Behçet disease: A case report

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Behçet disease is a multisystemic vasculitis of unknown origin. The vascular complications as a result of the disease are rare in the pediatric age group. We report a 13-year-old patient with vasculo–Behçet disease with a ruptured abdominal aortic aneurysm without a formerly known history of Behçet disease. Urgent aortoiliac bypass with a polytetrafluoroethylene graft was performed with success, and the patient has also received corticosteroid and immunosuppressive drug therapy. (J Vasc Surg 2004;39:901-2.)

Behçet is a clinical syndrome, initially defined by the presence of aphthous stomatitis, recurrent uveitis, and genital ulcers, that now is recognized as a multisystemic vasculitis with an undulating course of exacerbations and remissions.[1,2] There is no universally accepted diagnostic test for Behçet disease. We describe an unusual presentation of Behçet disease that has not been reported previously.

CASE REPORT

A 13-year-old girl was admitted to the emergency department with complaints of severe abdominal and back pain, progressive fatigue, and dizziness. At physical examination her abdomen was tender, and femoral artery pulses were absent. Blood pressure was 70/40 mm Hg, and cardiac auscultation was normal except for sinusal tachycardia. Chest x-ray films were normal. Laboratory investigations revealed a hemoglobin level of 8.2 g/dL and hematocrit of 21%. Bedside abdominal ultrasound revealed a retroperitoneal hematoma secondary to perforation of an infrarenal aortic aneurysm.

The patient was taken to the operating theater urgently. After median laparotomy, the large hematoma was seen in the retroperitoneal field. The proximal aorta below the renal arteries was clamped, and the aneurysm sac was opened. Massive fresh hematoma and blood were aspirated from the operative field, and the perforation site was seen on the left posterior wall of the distal abdominal aorta, including the proximal left common iliac artery. It was a true aneurysm of the distal abdominal aorta, approximately 3×4 cm in diameter. An intimal tear and 4-cm long ulceration of the left posterolateral aortic wall were associated with the rupture site. The continuity of the left common iliac artery was interrupted as a result of rupture of the aneurysm sac, which extended toward the external iliac artery. Abdominal aorta reconstruction was performed with a straight polytetrafluoroethylene (PTFE) 10-mm prosthesis, below the renal arteries to the orifice of the right common iliac artery. A separate 8-mm PTFE graft was used for reconstruction of the left common iliac artery, because its lumen could not be seen. The distal side of the graft was anastomosed end-to-end to the left external iliac artery, and the proximal side was anastomosed end-to-side to the main graft. After intensive care therapy for 5 days, rapid healing was observed, and the patient was transferred to the rheumatology clinic for diagnosis of the possible cause of the aortic rupture.

The patient has had complaints of arthralgia of the knees, oral aphthous lesions, anorexia, and weakness for the last 3 months. She also described a history of minor genital ulceration, but we did not find a lesion at physical examination. The pathergy reaction, a hypersensitivity of the skin to a needle prick, which is peculiar to this syndrome, was positive. Rheumatoid factor, anti-DNA, human leukocyte antigen–B5, and antinuclear antibody were normal. The erythrocyte sedimentation rate and C-reactive protein concentration were mildly elevated. Ocular involvement (uveitis) was not detected at fundoscopic examination. Pathologic study of aneurysmic aortic wall revealed subintimal infiltration of inflammatory cells, destruction of the lamina elastica interna and smooth muscle cells, and multiple necrotic regions on the tunica media (Fig). Mycotic aneurysm was ruled out with clinical findings and microbiologic cultures. In addition, computed tomography of the thorax was performed to determine whether the thoracic aorta was involved. The diagnosis of Behçet disease was made on the basis of the positive pathergy test, oral aphthae, short-term arthritis (a few weeks), and history of genital ulcers. Cyclophosphamide (15 mg/kg, administered in monthly intravenous boluses), methylprednisolone (0.2 mg/kg), and oral anticoagulation therapy were started; and after 3 months cyclophosphamide was changed to azathioprine (3 mg/kg/d). After the first month of therapy, the patient's symptoms markedly diminished.

DISCUSSION

Inflammatory abdominal aortic aneurysms are a distinct entity characterized by marked thickening of the aneurysm wall, which frequently leads to urgent exploration for suspected ruptured aneurysm, as in our patient. The clinical spectrum and severity of Behçet disease depends on extension of vasculitis. Because the disease affects blood vessels of different types and sizes, manifestations may occur at many body sites. The diagnosis of Behçet disease in children can be difficult because of the rarity of the condition and the nonspecific manifestations. Although Behçet disease is a multisystemic disorder, it may involve only a single organ during childhood, which could delay the diagnosis. As in

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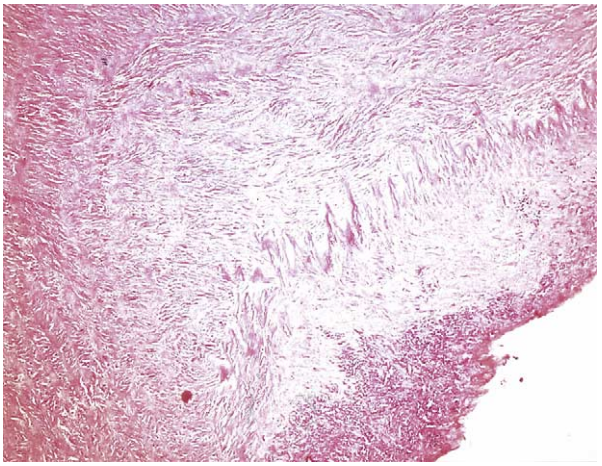

Photomicrograph demonstrates the typical pattern of destruction of the medial elastin, and prominent inflammatory infiltration with associated necrosis in the subintima and tunica media (hematoxylin-eosin, original magnification ×40).

our patient, some patients with Behçet disease fail to exhibit the classic symptoms and signs of genital ulcers and uveitis. A third of the patients of Kone-Paut et al[3] failed to fulfill the international criteria for the diagnosis of Behçet disease.

The classification of vasculitis is confusing, because there is considerable clinical overlap between the various vasculitic syndromes. The diagnosis of vasculitis requires histologic confirmation in most cases, and the classification is based on the size of the predominant involved vessels. In our patient we first thought that one of three probable vasculitis disorders could be responsible, that is, Ehler-Danlos syndrome, Behçet disease, or Takayasu disease. Ehler-Danlos syndrome is characterized by hyperextensible skin, hypermobile joints, fragile tissuses, and bleeding diathesis related to fragile vessels. Patients are prone to spontaneous rupture of major vessels, aneurysm formation, and acute aortic dissection,[4] as in our patient. However, both the absence of concordant clinical findings and presence of acute inflammation with associated necrosis at histologic section analysis exclude Ehler-Danlos syndrome. The absence of arterial occlusive symptoms and clinical findings exclude Takayasu disease, or pulseless disease.[5]

The pathogenesis of aneurysms in Behçet disease is multifactorial and quite different from that of atherosclerotic aneurysms. Abnormalities of neutrophils, enhanced adherence to endothelial cells, and increased adhesion molecules have been suggested as responsible in the pathophysiology of vascular involvement. Yamana et al[6] demonstrated a relationship between immune complex vasculitis and Behçet disease. They showed large deposits of immune complexes in the intima and surrounding tissue. These findings suggested that immunologic reactions occur on the inner and outer sides of the artery at the same time, leading to destruction of the media and formation of an aneurysm.

Vascular involvement in Behçet disease was reported to range between 7.7% to 60% in adult series,[7] but in pediatric patients arterial aneurysms and complications are rare.[3] Large arterial aneurysms, which are more common than occlusions, are a major cause of death, because of the risk for rupture.[8,9] Usually abdominal aortic aneurysms are discovered in the chronic stage, with vague symptoms of back pain and abdominal discomfort. In our patient rupture of the abdominal aorta to the retroperitoneal area was the first symptom of the aneurysm. Abdominal aortic aneurysms as a result of Behçet disease are usually seen in men; Tüzün et al[9] did not detect a single aneurysm in a woman, in 2179 patients.[9] Vascular complications did not develop for 3 to 16 years after disease onset.[7-9] Our patient is younger than 16 years and female, and onset of symptoms was less than 6 months before aortic rupture.

Predicting which patients will have cardiovascular complications is the major concern of recent investigations. To our knowledge, this is the first report of abdominal aortic rupture in a child as a result of Behçet disease. Early diagnosis of vascular involvement is helpful for planning effective management and improving the prognosis. Long-term follow-up is also essential in patients with Behçet disease, because of the relapsing nature of the disease.

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